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OF THE CHILDREN'S HOSPITAL

WASHINGTON, D. C.

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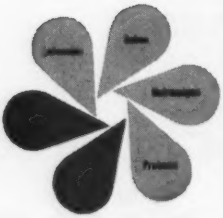

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EMOTIONAL ASPECTS OF A CHILD WITH POLIOMYELITIS

Gene Gordon, M.D.,* Belinda C. Straight, M.D.,† Mary Robinson, M.A.,‡ Joseph M. LoPresti, M.D.,§ and Emma Layman, Ph.D.‖

INTRODUCTION

Dr. Gordon:

When a child becomes sick, like the rest of us, he becomes frightened—frightened and I might say angry. For one thing, he is afraid of death, particularly when he senses that a disease like polio is possibly lethal, and he is, in any case, afraid of injury. Today's case presentation and the discussions which follow will demonstrate how such feelings are manifested by a 4 year old boy with poliomyelitis.

CASE REPORT

R. K. is a 4 year old colored boy admitted to Children's Hospital August 10, 1957 with a two hour history of frontal headache and anorexia, and a 24 hour history of fever. The patient was the product of a full term pregnancy. Birth weight was 7 pounds 5 ounces. There were no complications after birth. Measles was the only childhood disease. The patient previously had been admitted to Children's Hospital when he was 18 months old and was diagnosed as having follicular tonsillitis, nutritional anemia and diarrhea. He had had no poliomyelitis immunizations. His mother was 33 years old, a kitchen worker, living and well. His father was 44 years old, epileptic, and not employed. There are three siblings living and well and one 10 year old brother who had had rheumatic fever. They live in a house with two bedrooms. The father takes care of the children while the mother works.

On admission to the hospital the patient was lethargic and acutely ill, but fairly well hydrated. There was definite neck and back stiffness and a positive Kernig sign. There was no definite muscle tenderness. All reflexes were brisk and equal. Cerebrospinal fluid contained 244 cells per cu. mm. with 92 per cent polymorphonuclear cells and 8 per cent lymphocytes, 120 mg. per 100 ml. of sugar, and 60 mg. per 100 ml. of protein. Although the diagnosis was considered to be poliomyelitis, the patient was given antibiotics due to the possibility of bacterial meningitis. Three days after admission, he was still febrile in spite of antibiotics. At this time, the patient had definite neck stiffness, positive Kernig sign and absent knee and ankle

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jerks. On repeat examination the cerebrospinal fluid had 35 cells with 80 per cent lymphocytes. On the fifth day after admission the temperature returned to normal. On muscle evaluation there was noted a considerable hamstring spasm and tenderness with decided weakness in the lower extremities. On August 19, at the time of transfer to the convalescent poliomyelitis ward, the boy was moderately irritable and uncooperative and had hamstring and back spasm. A consultation with the Department of Psychiatry was requested because of episodes of inappropriate crying alternating with episodes of aggression and belligerence.

PANEL DISCUSSION

Dr. Straight:

The Department of Psychiatry was asked to see Robert because he was uncooperative. This behavior was disturbing to the others on the ward. At times he was withdrawn, at other times he was hostile. He fought against lying down in a polio jacket. He had temper tantrums when he could not get his needs met immediately. He soiled and wet and threw his underpants across the room. This behavior started soon after hospitalization.

From Robert's mother, who is a hard working, conscientious, warm woman in her middle thirties, we have the history of his previous adjustment. Robert used to be an easygoing child. He played well with his brothers and sisters, and could share in ordinary family life comfortably. Developmental history was normal except in two areas: one was that the mother went back to work within a month or two after his birth to support the family; the other was that his father and a friend took care of him. Toilet training occurred at two, but he has had occasional episodes of bed wetting since then. He walked and talked and socialized at the usual time.

The father of the family began to have a convulsive disorder five years ago and since that time has episodically drunk alcoholic beverages to excess. He has frequently been unemployed. While drinking and while sober he is easygoing, and is particularly close to the patient. Robert also has two older brothers, a younger one of 9 months, and an older sister. The sister and Robert are frightened by their father's convulsions which they have seen several times. They cry and cling to one another.

In my interview with Robert, he greeted me loudly. He would frequently call across the room to other adults. He is active, stocky and more concerned with direct action than with words. Sometimes he wrinkled up his nose as though things were distasteful to him. His affect was appropriate, his mood neutral. He wanted to make friends right away. He became disorganized if his needs were not met immediately. When asked how he got ill and what kind of illness he had, he answered that it was some trouble with his head. He is better now. He did have

some needles, but he did not have polio. As this did not get at very much material directly, we played with the doll family. We made one doll be a sick boy and had a doll doctor. Robert was interested in the dolls' legs and he handled them gingerly. When asked how the little boy got sick he replied, "Because the girl doll put a worm in his mouth. It has gone down inside of him and it broke his leg. If the boy keeps his mouth open, the worm will come out. Sometimes boys get sick from dirt." When asked if some of the dolls had polio, he quickly quipped "Let's play something else." He later added that doctors give needles to make people well, but sometimes they do this to make people sick.

In summary of my impressions, Robert displays anxiety over his hospitalization and illness, which shows in his regressive and explosive behavior. Much of this anxiety arises from his feelings of helplessness, which is emphasized when he is placed in a polio jacket, and in a prone position. We see the regression in his being unable to share, unable to wait for what he wants and his soiling and wetting. There is anger about the illness, both toward his family for bringing him to the hospital, and toward the doctors and nurses for endangering him in certain ways. We see that he uses the mechanism of denial about his illness although at the time when I saw him, he had been told about his polio. This defense of denial does not take care of the tremendous amount of anxiety he has about his own body, what his illness means or how he got it, including what bad things he thinks he did to make him sick.

We know that parents have much difficulty in telling their children that they have polio, and what polio means. Also, doctors often have difficulty telling their small patients, partly because they often do not know what the child has already been told, and partly because it arouses anxiety in the doctor. Occasionally, a child may be on the ward a few days before he hears his diagnosis. Such a child may feel that he has been fooled or tricked, and then it is harder for him to trust and cooperate in other aspects of his care. On the other hand, children who have been well prepared about their illness may use denial to stave off the uncertainties and anxieties arising from their hospitalization and illness.

Mrs. Robinson:

I did a psychological evaluation on Robert which was rather brief, but I think somewhat productive. He is a very well developed boy who was introduced to me on the ward. In this setting he was eager to relate and he talked at great length. He seemed quite happy to see me although he had never met me before. When I suggested leaving the ward and going to my office he did require some reassurance about what would happen, particularly in reference to getting a needle. However, he rather readily

accepted my reassurance that I was not the kind of doctor who gave needles. He continued to relate warmly and talked quite spontaneously. One interesting aspect of his relating to me was that he seemed quite eager for physical closeness which is characteristic of a much younger child. He asked if he could come to see me every day, and he seemed concerned about the other children I might see, especially girls. He asked about one particular girl on the ward, and said that he certainly hoped that I was not seeing her. This girl was brought up several times, and he seems to have a close but intensely rivalrous relationship with her. He also asked me at one point if performing on the test tasks would make him well. He said, "Will all this buttoning make me well?" I asked him why he thought it would and he said that he had been told that the way to get well was to do everything the doctors told you to do.

I gave him the Merrill Palmer scale of intelligence and he performed at a dull normal level, with a mental age of 4 years, 4 months, and an IQ of 87. Although he would cooperate initially on every task that I introduced, he was continually distracted by his own ideas. In the middle of performing a task, where time was a very important element, he would suddenly stop and say, "Do you live here?", "Do you go home at night?", "Where do you live?". Ultimately, I felt that this distractability did contribute to his failure on many subtests, so that free from some of these concerns he might have performed on an average level.

I also gave him one of the picture story tests, and in these stories he expressed a great concern with people being naked, and even perceived fully clothed people in the pictures as being undressed. He said that this meant that they were crazy, and he seemed a little embarrassed by this. He saw a picture of a boy and a girl and said, "This is a boy and his mother. She's mad at him. She don't want him." On another card which is usually seen as a father and son picture he said, "They are talking about danger stuff." I asked him what danger stuff was and he said, "Danger stuff is being bad." Periodically, while responding to these pictures he would come out with many of his own thoughts, particularly in respect to his illness. At one point he said, "I'm crazy. I'm sick in my head." When I asked him what happened when he got sick he said, "I was sitting on the porch, and my head began to hurt and I was sick. I didn't know why I was sick." I asked him if he knew now why he was sick. At this point he was extremely embarrassed, turned his head away and giggled, and for the first time refused to answer questions.

My over-all impression is that this is a child of at least dull normal intelligence, although certainly his emotional concerns are depressing his functioning in all areas. Emotionally, he presents a picture of a rather anxious child who is concerned about the reasons for his illness, and quite

confused about them. I think he feels that polio is a consequence of his own badness in some respect, that it is his fault, and that his hospitalization may represent to him a rejection on the part of his family. In judging the child's relationships, he does relate to adults, at least from my own experience, on a rather immature level of physical closeness. He is intensely competitive with other children, particularly with girls, in trying to relate to an adult figure. My impression was that this is not a severely disturbed child, but that he is having personality problems which probably have been accentuated by his illness.

Dr. Gordon:

A boy, 4 years of age, in general, under the best of circumstances, has not developed to the point where he is clear on many things, and is apt to think of things in a magical kind of way. He is not yet clear on what is real and what is not real; he is not clear on the difference between ideas and actions. He still has notions that he, through his thoughts and wishes and daydreams, can magically control the world. It is hard for most of us to appreciate just how much magic thinking does go on in the mind of a child of 4 or 5 years of age, and how hard it is for him sometimes to distinguish between his daydreams—and his night dreams for that matter—and what really happens in the world. I was struck by this recently, for example, in connection with the satellite. My own children seemed very unsurprised and were unaffected to a large extent. Why? Because for months and years on television, in stories and in comic strips, there has been space travel, and I am sure that most children of 5 or 6 years of age consider that space travel is already a fact. Similar thinking applies, then, when it comes to the fear and the anger which are generated by an illness. We have to understand the way that the child feels, and the way that his fears and aggressions fit into his life. The way in which he reacts to them has to do with what his current fantasy life is, or what his daydreams are like, as well as what the realities are. Now as I said, a boy of 4 or 5 years of age, under the best of circumstances has trouble differentiating between daydreams and reality, but part of the difficulty with our patient is due to what we call regression, which simply means that when threatened with an overwhelming physical insult or a threat to one's life, all of us tend to go back to earlier modes of functioning and become more childlike. This is true in the heat of battle, when brave and strong men will call out for mother; it is even truer in the case of a child whose character and personality are not yet set. One aspect of this going back to earlier kinds of reactions is that he becomes even less capable than under normal circumstances of differentiating between fact and fancy. His fantasies play a disproportionate part in his reactions to the world.

In Robert we have some clues as to how he handles his fears and his anger. He, like many children and many adults, particularly adults with very severe illnesses, reacts first by what we call denial. That is, he says, in effect, that the threat does not exist, "I am perfectly well." Interestingly enough, the worse the illness the greater the need for the person, adult or child, to deny its existence. To a certain extent all of us deny the existence of death all the time; when we stop to think about dying we tend to get kind of depressed or upset. I think most of us have deep down a kind of conviction that we are going to live forever. This is even more so in people who are imminently threatened by death. In a study at the Massachusetts General Hospital, for instance, it was found, curiously enough, that the respirator cases and those with bulbar polio were very much more cheerful and happy than the milder paralytic and non-bulbar cases. Why was this so? Because the respirator cases, being threatened more imminently, had to react with an even more total suppression of inner feeling. I think the denial which is going on in this boy is pretty clear. He is not sick now. He used to be sick, and the sickness was in his head, and no other part of him was involved. I was interested in his saying that no one visits him except his cousin, although his parents have been visiting him faithfully. This might be a reflection of his anger with them, as Mrs. Robinson suggested, conceivably for abandoning him by having him hospitalized. Given the fact that the fear and the anger engendered by the illness are made part of the child's fantasy, this has several implications. Depending on what his daydreams have been, he is going to distribute this anger, so to speak, in the environment. That is, he is going to blame himself in some instances, and feel guilty as this boy apparently does. He tells us that he ate something he should not have eaten and this is what made him ill, or that he played in the mud when he should not have, and this is what made him ill. Or his anger may be directed to outside agencies, such as his parents who have made him ill. He may also turn the anger against himself if he is afraid of his angry feelings. He then gets quite depressed or moody. In the extreme, the depression may go even to the point of suicide through starvation. More hopefully he may diffusely discard the anger all over the place, which seems to be this boy's pattern. We have heard of his temper tantrums, messiness, and so on. He may, as many patients do, adult patients particularly, react in the opposite direction, saying in effect, "I'm not angry. I love everybody." Such patients become very helpful to the other patients. They become very concerned about other people whom they consider to be worse off than themselves. Incidentally, this notion of others being worse off than themselves may have little or no relation to reality, but may be another part of the denial.

Sometimes the child can find substitute modes of expressing the aggression in his play activities or in daydreams, if direct expression in action is too dangerous or frightening. People who are chronically sick with a disabling disease like this develop, of course, many compensatory modes of chanelling their aggressions. One can use the aggressive feelings in constructive ways as well, such as in building things in occupational therapy. It is interesting, incidentally, that the immobilization in this particular disease, the fact that the child is not able to move, accentuates the problem a great deal. Characteristically we express anger in movement, either diffuse tantrum-like movements, or striking out, or more controlled and chanelled movements, such as athletics. For children, deprivation of movement is an extreme punishment and poses a great threat to them, because it is such a useful way of disposing of feeling. Very often we see, then, two possible choices for the child who has paralysis or some other immobilization. One is that the inhibition may spread, so to speak, and involve other parts of his body. Speech, for instance, or visual functions can become impaired, or, (and this, of course, is a more hopeful solution for it) these other functions can be used to excess. There is the story of a little girl who, unable to strike out or move, paid her friends to bring her dirty words. For each dirty word she would pay so much out of her allowance. Thus she was able to use speech in a compensatory way to express some of the anger which she could not otherwise do because of the immobilization of her body. There is a great deal more that one could say about polio generally, and Robert in particular, but I would like to leave a little time for questions and discussion.

Dr. LoPresti:

It is interesting to note that this year we are seeing a changing trend in polio. In the past, polio was primarily a disease of children from a fairly good environment, economically stable with fairly intelligent parents whom you could approach and talk to and explain things. The change in trend is that 98 per cent of our patients this year are colored children who come from overcrowded homes and whose parents are not as intellectually mature as the parents that we have seen previously. This makes the way in which we approach parents a different problem entirely. I would like to give you a little picture of what happens to one of these parents when he comes in. A parent comes in with a child who is sick. The resident physician in getting his history talks to the parent, and one of the questions he asks, if he suspects polio is, "Has your child been vaccinated against polio?" The parent says "No." The first thing that the resident physician says is "Why not?" This immediately tends to place the blame for the whole thing on the parent. If there was no guilt to begin with he

has planted the seed for it. If there was guilt there to begin with he has accentuated it. The nature of this disease makes it very difficult for us to talk to parents initially and explain what is going on. When we see these children in the acute stage, there is no way of predicting while the child has fever whether paralysis is going to develop, how severe the paralysis is going to be, or whether, indeed, the child may not even die. This creates a waiting period of about a week of severe anxiety on the part of the parents, and on the part of the child if he is old enough to know what is going on. The other thing that I think you, as a psychiatrist, should be concerned with in this particular patient is the child's outlook. According to his chart he has absolutely no function of his anterior tibial and peroneal muscles, and after three muscle evaluations there has been no improvement. It would appear safe to assume, therefore, that there will be no improvement in these paralyzed muscles of his left leg. I can predict that he will be hospitalized for a minimum period of six months, and probably closer to a year. If he thinks he is not sick, how are we going to explain that he is going to be here for a year? In addition to that, I think you should be fully aware of the probability that ultimately this child will require some form of orthopedic surgery.

Dr. Gordon:

I think you raised some very interesting points. Certainly there is a feeling of anger on the part of the physician toward the parents because they have not arranged that the child be immunized. With any disease in which we are helpless we feel pretty angry. I think not only is it the fact that we cannot predict the extent of the paralysis in the acute stage that makes us anxious, but the fact that we cannot do anything about it either, except perhaps administer only the most symptomatic treatment. This in turn puts us in almost the same helpless position as the child. Being helpless we get angry, not to mention the fact that part of the anger of the child and the parent is directed toward us, and this I think we perceive. When there is something that they could have done, it gives us something to hang our anger on. What is bad about this is that it increases the parents' guilt, and they in turn take it out on the child and back out on us, and we have in the long run a less cooperative parent and a less cooperative child. Therefore, if we could perhaps try to understand some of the helplessness and hopelessness of these people that has kept them from organizing their lives the way we think lives ought to be organized, it might make it a little easier for us to be sympathetic toward them in their dilemma, which could then in turn make them more cooperative.

As far as the child is concerned I think we do have to bring him to some recognition of the realities of his illness. One way to begin this is to deal with it realistically ourselves from the beginning and not contribute, so to

speak, to the denial which the child and the parents seek by leaving it a total blank. I do not know that polio or any other diagnostic term is very meaningful to a child. I think that with time and effort spent with the children to find out what their own notions are about this disease we can help, working from their own notions, to bring them closer to what the realities are. This particular child obviously has a wealth of distortions about what the illness is, and one would have to take up these distortions and clarify them one by one. It is not enough simply to diagnose the disease for him, or to give a description of the illness. That is only part of it. The other part is to try to understand what his own distortions are so that you can correct them; and the distortions are manifold. Anything you can conjure in your wildest imagination, some child can imagine and believe about his illness. Also, you are going to try to help him find ways of chanelling the aggressive feelings that are mobilized by the illness, so that they can be discharged, and discharged without jeopardizing his role in the ward society, alienating the nurses, the doctors and the other children. Again, it is only through spending time and developing a personal relationship with the child that one can help him in these ways. Our occupational therapy program is one way of doing this. Before you can help him to develop you also have to allay his anxieties, particularly during the acute phase. Because the anxieties are exaggerated by his regression to infancy and his great dependency on the people around him one has to go rather gently with him in order to supply him with strong, stable, persistent relationships. That will give him confidence in us and will enable him to transfer to us some of the confidence, hopefully, that he has in his parents. Of course, how much confidence this child has in his parents from the brief history we had, we do not know. The mother is described as a rather conscientious person.

Dr. Layman:

It would seem to me that an important aspect of the doctor's handling of the situation is his dealings with the parents and the child. One has to be honest and forthright. It is very misleading and confusing to give the impression that the child will be the same as before his illness. For example, statements such as "You will be home in no time at all," or "You will soon be playing football," can only lead to distrust and loss of confidence in the physician. Parents may be guilty of this even when the physician is not.

Dr. Gordon:

We tend to want to soothe both the parents and the child. This, in the long run, only destroys trust and makes it more difficult for a child to grow back up.

Dr. Straight:

The children whom I have seen in the respirator were usually children whose mood was rather contented, who accepted it rather easily and whose general spirits of well being were quite high. I wonder if this is characteristic.

Dr. LoPresti:

Before we place a child in the respirator one of the things that we have done up on the ward is to bring the respirator in, sit down and spend a good deal of time with the child explaining what the respirator is, why it is being used, and that as soon as he is able he will start coming out of it. I think the preparation of the child is very important to success.

Dr. Gordon:

Is it true that the anxiety about going into the respirator is often much less than that about coming out?

Dr. LoPresti:

Yes, that is true. All the time that they are in the respirator we keep stressing that we are going to get them out as soon as possible. This keeps the child thinking, "Well, I want to get out of this thing", and we have been quite successful.

Dr. Gordon:

The respirator has assumed a tremendous importance in the life of the child. For one thing, another part of the regression, if you will, is that the interest of the patient, of any sick patient, is withdrawn from the rest of the world, so to speak, and is concentrated on those aspects of his life on which his life depends: on the nurses, the doctors, the parents, and in the case of the respirator patients, the apparatus itself. The piece of equipment itself becomes as important to him as his mother was in the first few months of life. One report of the Boston epidemic mentioned that there were lots of hurricanes at the time and the patients were extremely anxious reading or hearing about them, because hurricanes meant electric power failure, which in turn meant respirator failure. The whole world was condensed into a respirator for these patients. One has to be aware of this in helping the child to make the transition out of the respirator. Rocking beds are a great help in such transitions.

AORTIC ARCH ANOMALIES

George William Ware, M.D.,* and Robert T. Scanlon, M.D.†

HISTORY[6, 12, 13, 14]

Surgery for anomalies of the aortic arch had its inception in 1945. However, descriptions of these entities date back to the eighteenth century. Hammell, in 1737, described the first case which happened to be a double aortic arch. Bayford, in 1794, reported an aberrant right subclavian artery. He noted that it originated from the descending aorta below the origin of the left subclavian artery, and that it passed upward and to the right, compressing the esophagus in its passage. Bayford labeled the condition "dysphagia lusoria" or esophageal obstruction due to a freak of nature.

Such conditions, however, were autopsy room curiosities until Gross introduced surgical treatment. In August 1945, he operated on a 7 month old infant with a double aortic arch and divided the smaller anterior limb. In November of the same year, he resected an aberrant right subclavian artery which arose from the descending aorta. Due to these pioneer efforts, surgery is now the accepted treatment for symptomatic cases of aortic arch anomalies.

EMBRYOLOGY[1, 4, 10, 11, 13, 16, 18, 20, 22]

It is our intention to describe only the more common symptomatic varieties, namely:

1) Double aortic arch
2) Right aortic arch with left ligamentum arteriosum
3) Aberrant right subclavian artery
4) Anomalous innominate or left common carotid arteries

More complete classifications are available in articles by Edwards, and Gross and Ware.

To understand these entities, some knowledge of the basic embryology is essential. During the first few weeks of embryonic life, six aortic arches appear. Their function is to join the ventral aortic sac or bulb with the dorsal paired aortae. The first two arches quickly disappear. The third aortic arch becomes the proximal portions of the common carotid arteries. The left fourth arch presents as the normal future aortic arch. The proximal portion of the right fourth arch becomes the innominate and right subclavian arteries. The distal portion of this right fourth arch normally becomes obliterated. (The left subclavian artery arises from the sixth

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cervical intersegmental artery.) The fifth arches disappear without a permanent role. The ductus arteriosus is formed from the sixth left arch.

With this brief description, the usual anomalies should become apparent. If both fourth aortic arches persist, a double aortic arch is present. A right aortic arch occurs when the left fourth arch becomes obliterated instead of the right fourth arch.

If the proximal portion of the right fourth arch disappears rather than the distal portion, the right subclavian will arise distally on the descending aorta.

SYMPTOMS[6, 7, 11, 14, 16, 18, 19, 22]

All varieties may occur without symptoms. The degree and extent of compression of trachea and esophagus are the major factors. If there is no compression of the lumens of these organs, there are no symptoms.

Double Aortic Arch. Difficulty occurs early in life and may be noted immediately after birth. Respiratory symptoms predominate. Audible breathing is the rule, Wheezing, inspiratory and/or expiratory stridor, and dyspnea on exertion are common. The infant nurses slowly and often becomes cyanotic during feeding. Repeated respiratory infections and pneumonias are likewise common.

Right Aortic Arch with Left Ligamentum Arteriosum. Symptoms with this entity occur in childhood rather than early infancy. Frequent respiratory infections occur. The respirations are often described as "crowing." Dysphagia is not a prominent complaint.

Aberrant Right Subclavian Artery. In this instance, esophageal complaints are the more prominent. Dysphagia or hesitancy in swallowing is seen. The onset of symptoms is later than with a double aortic arch since liquids are often ingested without difficulty. When solid foods are added to the diet, the resultant dysphagia is such that the child refuses to eat. If feedings are forced, vomiting and aspiration may occur. The mother usually learns that the patient does best on small frequent feedings.

Palmer has reported a series of symptomatic cases in adults. Dysphagia was the most prominent and often the only symptom. For some unexplained reason, most of the group dated onset of symptoms to the third decade of life. The senior author has recently treated an adult with marked compression of the esophagus and trachea due to this entity, yet this individual was asymptomatic.

Anomalous Innominate or Left Common Carotid Arteries. Symptoms are not common in this group. When they occur, they are respiratory in nature due to anterior compression of the trachea. Dyspnea, wheezing and stridor may be present. There is no esophageal compression, hence no dysphagia is present.

RADIOLOGIC ASPECTS[2, 3, 5, 6, 11-13, 15, 17, 21, 24, 25]

Radiologic study is usually diagnostic. The survey chest x-ray may be normal. However, with a right aortic arch, the aortic knob may be outlined on the right. On the lateral view, the trachea should be examined carefully for narrowing or anterior displacement. Laminograms will clearly define tracheal narrowing.

Fluoroscopic study is essential. Thick barium is preferred unless there is danger of aspiration, in which case iodized oil is then the preferred medium. In patients with a double aortic arch, there is a broad and deep posterior compression of the esophagus at the level of the third or fourth thoracic vertebra on lateral view. In the anteroposterior projection the esophagus is compressed from both sides. In patients with a right aortic arch and left ligamentum arteriosum, there is a narrow but deep impression on the left lateral and posterior walls of the esophagus. With an aberrant right subclavian artery, the lateral esophagram reveals a narrow indentation at the level of the third or fourth thoracic vertebra. In the anteroposterior view this line will often be noted to run obliquely. The trachea is of normal caliber except in those rare instances when the vessel runs between esophagus and trachea rather than posterior to the esophagus.

Angiocardiograms may be used, but usually are not necessary for the diagnosis. Raphael and associates have described a unique method of diagnosis of an aberrant right subclavian artery. They have passed a catheter into the vessel on the right, threaded it back to its origin from the aorta and then injected the dye.

ENDOSCOPY[10, 11, 19, 24]

Bronchoscopy will usually reveal a pulsatile compression of the trachea just above the carina. This compression is more often anterior in location. Esophagoscopy demonstrates a similar but posterior compression of the esophagus.

SURGERY[5, 9, 11-14, 22, 24]

Surgical treatment is necessary for all symptomatic cases. The operation is not without hazard. Difficult and meticulous dissection of major vessels is necessary in chronically ill, often emaciated infants. The operative risk is illustrated by Gross's statistics of five deaths in a series of 21 cases with double aortic arch.

A left thoracotomy is employed for all the anomalies described. Gross prefers an anterolateral incision. However, the standard posterolateral thoracotomy incision would seem preferable for wider exposure. The

thymus may be excised for more complete dissection. The surgeon, utilizing intimate anatomical knowledge, must expose all structures before dividing any part. He must keep in mind that a posterior arch may be difficult to demonstrate.

In the case of a double aortic arch, the smaller limb is divided. This is usually the anterior component. If the case is one of a right aortic arch with a left ligamentum arteriosum, only the ligamentum is divided. With an anomalous innominate or left common carotid, the condition is alleviated by suspending the vessel anteriorly to the sternum. A right aberrant subclavian artery is divided at the level of origin from the aortic arch.

COMPLICATIONS

Pneumonitis and malnutrition have already been mentioned as the more frequent complications. Paralysis of recurrent laryngeal and phrenic nerves may occur during dissection or from excessive traction on these structures. Permanent tracheal deformity is seen in some individuals due to prolonged compression and interference with growth of tracheal cartilage. Operative hemorrhage is an ever present danger. Postoperative respiratory obstruction is common and may be due to two factors: laryngeal edema subsequent to endotracheal intubation during prolonged surgery, and trauma to the trachea during surgical dissection.

CASE REPORT

C. L., a 4 month old white infant, was admitted to Children's Hospital on March 30, 1956, with a chief complaint of labored breathing for five days. History revealed that the patient was delivered by Caesarean section during the seventh month of pregnancy. Because of respiratory difficulty after delivery, the infant was kept in an incubator for 21 days. Breathing was always a noisy process and thick mucus production complicated feedings. The present illness began with respiratory difficulty, cough and fever. On the day of admission, the respirations were reported as gasping in nature.

On admission, the infant had respiratory distress with substernal retraction. Bilateral coarse rhonchi were heard in the lungs. There was a slight leukocytosis. A first-strength PPD was negative. X-ray examination of the lungs showed evidence of a right upper lobe pneumonia. Initial treatment consisted of oxygen, moisture and antibiotics.

The patient was seen in consultation on April 9. Diagnostic possibilities suggested were tracheo-esophageal fistula without atresia of the esophagus, dysphagia lusoria, and congenital laryngeal stridor. A barium swallow showed the esophagus to be markedly compressed and deviated to the right at the level of T3 and T4. The trachea was displaced to the right but not compressed (figs. 1 and 2). On April 12, esophagoscopy demonstrated posterior compression of the upper one-third of the esophagus; at the same examination the trachea was noted to be compressed anteriorly. The tracheal lumen was one half of the normal diameter. The infant

FIG. 1. Oblique view of barium swallow showing compression of esophagus

was seen by a cardiac consultant who agreed with the diagnosis of an aortic arch anomaly. The infant was discharged from the hospital to allow him to recover completely from his pneumonia.

He was re-admitted as an emergency on April 19, because of difficult and noisy respirations. Weight on this admission was 14 pounds. A left thoracotomy was performed on April 24. Dissection revealed an anterior aortic arch with innominate, carotid and subclavian arteries in normal position. Immediately posterior to the origin of the left subclavian artery the aorta seemed to buckle, pass posterior to the esophagus, and descend on the right side. The ductus arteriosus joined the aorta at this level. The anatomical findings were erroneously assumed to represent a left aortic arch with a right descending aorta. Since the ductus arteriosus seemed to be causing compression of the esophagus, it was divided.

Postoperatively the left leaf of the diaphragm was noted to be high in position; this was attributed to paralysis of the left phrenic nerve, although during surgery the nerve was carefully identified and protected. The infant appeared definitely improved following surgery, but continued to eat rather poorly. He was discharged on May 3.

Emergency re-admission was necessary on May 15. By history, the patient was well until May 10. On that date, fever, cough and noisy respirations appeared and became progressively more severe. Examination revealed a cyanotic infant in se-

FIG. 2. Oblique view of barium swallow showing compression of esophagus

vere respiratory distress who had sternal retractions. The lungs were clear except for bilateral rhonchi. Rectal temperature was 100°F. X-ray examination at this time revealed a left lower lobe pneumonia which was treated with antibiotics. The patient's symptoms were alleviated, but audible respirations continued, feeding was a serious problem and the patient lost weight. As his condition deteriorated, the logical question arose concerning the validity of the findings at initial surgery. An angiocardiogram was attempted but was unsuccessful.

Because of progression of symptoms, re-operation was deemed advisable, and performed on May 28. Marked adhesions were present and made dissection difficult. The entire arch, its branches, and the left pulmonary artery were carefully identified. When this was done, the true abnormality of a double aortic arch became apparent. The smaller anterior arch was then divided between the subclavian artery and the descending aorta.

The postoperative course was stormy. The patient remained dyspneic and cyanotic. Bilateral moist rales were heard in the lungs which, however, seemed to be well expanded. Feedings were refused or regurgitated. On the fourth postoperative day the patient received 80 ml. of salt-free albumin intravenously. Obvious pulmonary edema immediately occurred, necessitating rapid digitalization, the administration of positive pressure oxygen, and the application of tourniquets. The patient's condition was precarious for two days, after which he slowly improved.

The problem of adequate nutrition became foremost and gavage feedings were initiated. The patient slowly improved and gavage feedings were discontinued. He was discharged on June 27 when he was taking six ounces of formula per feeding.

The patient was again admitted to the hospital on October 5 because of fever, cough and respiratory distress. Chest x-ray failed to reveal any evidence of pneumonia. The left diaphragm was noted to be still high in position but motion was improved over the previous examination. Symptoms subsided after several days of therapy with antibiotics, and the patient was discharged.

A year later, on October 17, 1957, the mother wrote, "C is fine and walks and talks well. He will be two in December and is a perfect 2 year old."

SUMMARY

Aortic arch anomalies are an uncommon cause of respiratory distress and/or feeding difficulty in infancy. Symptoms, when present, may be diagnostic. Radiology is the most valuable means of diagnosis; x-ray views in the anteroposterior and lateral projections are essential. Endoscopy may be of value in confirming the diagnosis. In the symptomatic cases, surgery is essential for cure.

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CLINICAL PATHOLOGICAL CONFERENCE

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CASE REPORT

This 2 year, 9 month old white boy had been fairly well until November 28, 1956, when he had diarrhea and high fever accompanied by intermittent vomiting. The fever was controlled with the administration of aspirin tablets. On the following day, shallow breathing developed and the pulse rate increased, but the vomiting subsided. On November 30, because of persistent fever and diarrhea, and a moderate stiffness of neck, the child was admitted to Children's Hospital.

Past and family history were not contributory.

On admission, the child was well nourished, in no distress and weighed 32 pounds. Temperature was 102.6°F., pulse rate 136 per minute, and respirations 30 per minute. A moderate nuchal rigidity was noted. The remainder of the physical examination was normal.

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The following initial hemogram was obtained: hemoglobin 13.1 Gm. per 100 ml., volume of packed cells 44 per cent, leukocytes 7,400 per cu. mm., with segmented cells 35 per cent, bands 31 per cent, lymphocytes 23 per cent, monocytes 8 per cent, eosinophiles 3 per cent, platelets adequate. The urinalysis showed a 3 plus acetone reaction, with a pH of 5.5. Carbon dioxide combining power was 17 volumes per 100 ml. A pathogenic strain of *Escherichia coli* was cultured from the feces. The child was given an intravenous infusion of 350 ml. of ⅙ Molar sodium lactate followed by 500 ml. of 5 per cent glucose in water with 1 ampoule of Berocca C.® added, and 350 ml. of Travamine®. He also received chloramphenicol and a clear fluid diet. During the administration of an enema he was noted to have a short period of cyanosis. On the day following admission, neomycin was given by mouth and the oral intake was restricted to tea and water. During the day, the patient vomited a moderate amount of coffee ground material. By December 2 (the third day of hospitalization), the diarrhea subsided but the child vomited a moderate amount of green material. On the following day, he was offered a 1 to 3 dilution of protein milk along with tea, Lytren® and water *ad libitum*. He was sleepy all day and by night refused all fluids. In the early morning of December 4, he vomited a dark green material and passed a dark green stool with a moderate amount of bright red and dark clotted blood. Later during the day, he passed liquid bloody stools twice.

The following hemogram was obtained: hemoglobin 10.5 Gm. per 100 ml., volume of packed cells 34 per cent, leukocytes 17,500 per cu. mm., with segmented cells 66 per cent, lymphocytes 26 per cent, monocytes 8 per cent, platelets adequate. A barium enema was performed and no abnormality was reported; the patient was transfused with 300 ml. of whole blood. During the night, the child passed several bloody stools; these became more frequent the next morning. He then vomited bright red blood and dark clotted blood. At this time, the prothrombin time was 53 per cent of normal. The hemoglobin was 6.8 Gm. per 100 ml. and volume of packed cells 20 per cent. An upper gastrointestinal series was reported to be normal. After transfusion with 500 ml. of whole blood, within 10 hours the hemoglobin was 8 Gm. per 100 ml. and volume of packed cells 27 per cent. The Lee-White clotting time was three minutes. The hemorrhagic process continued throughout the night despite measures undertaken for the management of a possible bleeding peptic ulcer: Demerol®, Amphogel®, belladonna, Meulengracht diet and Mephyton®.

A laparotomy was done. The external aspect of the entire gastrointestinal tract and other abdominal viscera was examined and was found to be normal. The abdomen was closed and the child was returned to the ward in satisfactory condition. A Levine tube was inserted in the stomach and Wangensteen suction was started; this produced an intermittent return of coffee ground material. No other bleeding manifestations were observed for the next 10 hours following surgery. Profuse hematemesis alternating with profuse defecation of blood recurred in the early morning of December 9. After 48 hours of constant hemorrhage during which 3,500 ml. of whole blood were transfused, the child expired on December 11, 14 days after the onset of the first signs of illness.

DISCUSSION

Dr. Brown:

This patient presents the problem of an acute gastrointestinal infection complicated by dehydration, acidosis and later, by gastrointestinal bleeding. The diagnosis of a gastrointestinal infection due to pathogenic *Esch-*

erichia coli was established, and proper antibiotic therapy was instituted together with therapy for the dehydration and acidosis. However, everything did not go as well as expected, for later, he began to show evidence of bleeding from the gastrointestinal tract. The first evidence of bleeding was from the upper gastrointestinal tract, that is, there was coffee ground vomitus; this later was followed by melena and hematochezia. In other words, we have bleeding from the gastrointestinal tract manifesting itself in all three ways, by hematemesis, hematochezia and melena. Such manifestations make it difficult for us to localize the site of the lesion. Certainly, in an individual who has all three of these, we think primarily of something high up in the gastrointestinal tract. However, oftentimes gastrointestinal bleeding is associated with blood dyscrasias. There is no evidence in our protocol that indicates that any of these blood dyscrasias existed. I mention them primarily for consideration as possible causes of gastrointestinal bleeding. We would include in this group leukemia, thrombocytic and non-thrombocytic purpura, hemophilia, and hemolytic and aplastic anemias. There may be others which I have not included. I am inclined to think from the protocol that our lesion is not something extra-alimentary, but rather something in the gastrointestinal tract itself.

Turning, therefore, to the alimentary tract proper, we divide the lesions into those of the lower and upper tract. In the lower gastrointestinal tract, I shall mention several causes of bleeding, some of which we should consider and some of which we should not. Intussusception is a frequent cause of gastrointestinal bleeding and manifests itself by bleeding per rectum. I do not recall having heard of a case of intussusception manifesting itself by hematemesis. Also, one would expect to have felt a mass on abdominal examination even before exploration, as well as evidence of the intussusception at operation. Polyps of the colon and rectum are frequent causes of bleeding, but these cause rather intermittent bleeding episodes without hematemesis and usually are not a cause of exsanguinating hemorrhage. It is entirely possible for a large polyp to erode and bleed to the point of exsanguination, but our barium enema would very likely have demonstrated it if this were the case. Meckel's diverticulum has been reported to be the cause of severe gastrointestinal hemorrhage and this usually is the case when there is gastric mucosa in the diverticulum or the adjacent small bowel; this may be a cause of ulceration and hemorrhage. Certainly, anorectal conditions which are frequent causes of bleeding could not produce the picture that we see here. Multiple hemangiomata of the intestinal tract, though extremely rare, can be the cause of exsanguinating hemorrhage. If such were the case, we would have to presume hemangiomata to be present high in the small bowel, in the duodenum, or the jejunum, as well as in the large bowel and distal small bowel. In addition, with the hemangiomata, there are frequently other

noticeable superficial hemangiomata present. This child had no evidence of any skin involvement with this type of tumor. So then, we leave the lower gastrointestinal tract and come to the area in which all of these symptoms could arise.

In the upper gastrointestinal tract, we find esophageal varices occurring in children just as they do in adults. How would we rule out esophageal varices? The upper gastrointestinal series could show esophageal varices. However, in a child this young—2 years 9 months—the veins might readily be compressed by the barium as it passed through the esophagus and, therefore, might not be demonstrated. Esophageal varices are often associated with cirrhosis of the liver or with extrahepatic portal hypertension. Our abdominal explorations reported all viscera to be normal. Therefore, we assume no cirrhosis of the liver existed.

Peptic ulcers occur in children. They behave very similarly to peptic ulcers in adults; they perforate and they obstruct. Many times, in infants as well as in adults, severe gastrointestinal bleeding from peptic ulcers can go on to the point of exsanguination without ever being demonstrated at surgery or post-mortem examination, particularly if the peptic ulcer is in the duodenum. The diagnosis in this case might very well be that of hemorrhage secondary to a bleeding peptic ulcer. The first evidence of bleeding mentioned was the vomiting of coffee ground material which is indicative of blood having been in the stomach long enough to permit partial digestion. Peptic ulcer, I am told, is difficult to demonstrate by x-ray in young children.

Hiatus hernia could have been present, and if there were ulceration, bleeding could have resulted in the same sort of picture seen in a bleeding peptic ulcer. However, in hiatus hernia one would expect the greatest amount of blood loss to be by hematemesis. On the other hand, in peptic ulcer located in the duodenum where there is active peristalsis, blood can be carried rapidly to the lower gastrointestinal tract and be passed as free blood, or as blood that has undergone some change by the digestive process and appear as a dark tarry stool. Acute gastritis from poisoning or from acute infection could produce ulceration and bleeding from the stomach, but again, we would expect the greatest amount of bleeding to be by way of hematemesis. I include in the differential diagnosis small intestinal polyps which might lie high in the intestinal tract in the duodenum or the jejunum because these, too, can bleed very actively; the blood can be regurgitated as well as passed in tarry stools. Now, on abdominal exploration by our surgeons, a peptic ulcer could have existed and yet not be palpated or otherwise be demonstrable. Likewise, small intestinal polyps could be present and not be palpated through the intestinal wall, because they are so frequently soft.

Lastly, I have grouped together a few generalized conditions of the

gastrointestinal tract. First, an acute enterocolitis with diarrhea which was present in this child when he first became ill, and then progressed to ulceration with hemorrhage. Ulcerative colitis, unless it ascended into the small bowel, probably would not produce this picture. The swallowing of multiple metallic or glass foreign bodies, I assume, could produce excessive bleeding by laceration and erosion of the major mucosal or submucosal blood vessels. Recently, with the use of intensive antibiotic therapy, we have encountered staphylococcic gastrointestinal infections that have been rather severe and attended by a relatively high mortality rate. An acute enteritis produced by hemolytic *Staphylococcus aureus* can be fulminating, and although hemorrhage can be a part of the picture, it is not necessarily the cause of death. Toxemia is said to be the primary cause of death in this condition. Lastly, I mention typhoid fever. Typhoid fever can produce ulceration in the intestine and hemorrhage can occur secondarily. However, hemorrhage from typhoid fever would be expected to be found only expelled through the rectum.

After reviewing the differential diagnosis and rereading the protocol, I had at first felt that the most likely diagnosis in this child was an acute enteritis for which he was treated, with a complicating peptic ulcer which eventually led to exsanguination. I went back and thought a little bit more carefully about the possibility of the hemolytic *Staphylococcus aureus* infection involving not only the colon, but the small intestine as well. This could be the case, but my leaning is more toward high gastrointestinal ulceration. Therefore, my first diagnosis is a bleeding peptic ulcer and secondly, an acute fulminating enterocolitis with hemorrhage.

Dr. Guin:

Autopsy confirmed the fact that the child died because of exsanguination. There were 500 ml. of blood in the stomach and 500 ml. in the rest of the gastrointestinal tract. In the first portion of the duodenum, there was an ulcer which measured 2.5 cm. by 1.0 cm. which had eroded into one of the underlying major vessels.

The illustration (fig. 1) shows the duodenum denuded of the epithelium and the submucosa infiltrated with inflammatory cells. We can also see a branch of the pancreatico-duodenal artery which could have given rise to the hemorrhage. The absence of fibrous scarring is consistent with the short history of this patient's complaints. The pancreas was perfectly normal. The spleen showed extramedullary hematopoiesis, which was, no doubt, an attempt to compensate for the massive blood loss. The etiology of this ulcer is perplexing. When the child was first seen at home, the private physician was struck with the presence of the nuchal rigidity which disappeared after the child came into the hospital. She first enter-

FIG. 1. Area of ulceration in duodenum showing absence of epithelium and infiltration of inflammatory cells in the submucosa. A branch of the pancreaticoduodenal artery noted in the left of the field may have given rise to the hemorrhage.

tained the diagnosis of a meningitis and rightly so, according to the symptoms. However, the subsequent history did not bear that out. Sections from all parts of the brain stem were studied in an attempt to rule out an encephalitic component. We are currently interested in Cushing-Rokitansky ulcers because we see them so frequently. Frequently, cases at autopsy show ulceration of the upper gastrointestinal tract of the Cushing-Rokitansky type in known cases of central nervous system disease. For that reason we examined many extra sections of the brain of this child, but no pathology was found.

Dr. Straight:

This case of a child with a peptic ulcer was certainly a fulminating one. Sometimes cases of a more chronic nature are referred to the Psychiatry Department when the surgeon or pediatrician suspects family tensions as playing a role. Often we do not get to see them. Doctors frequently seem willing to agree that tension may cause ulcers in adults, but the idea that tension may have something to do with the causation of ulcers in children is harder to accept. However, we would like to suggest that, as part of the

diagnostic work-up, a psychological examination be given to determine whether the child does have considerable anxiety and other evidence of emotional disturbance. If the child does not have much anxiety, it would be thought that psychotherapy would not be of much value. Taboroff and Brown, at the University of Utah, studied children and adolescents with peptic ulcers. They believe that many of these children have had stressful life situations at home. Many of the parents of such children show immaturity and instability. Many of the mothers are directly rejecting women who find it difficult to relate warmly to their children or who are overtly overprotective. In other words, they may be overprotective in some ways but also push their children away in other ways, and the child feels somewhat caught between the two situations. It was found that children with ulcers often had fathers who were either absent from the home or were rather aloof from the family situation. Some of the children who were noted to have marked body tension seemed outwardly fairly passive. They did not engage in pushing other children around and in other active interests, but they did seem to have intense underlying and unexpected aggressions. They were overly dependent on their mothers, and at the same time had marked aggressive feelings toward them. Many of these children were shy. We also know that there are many patients who have this kind of personality makeup and who never do develop peptic ulcers. We might raise the question: why do some children who have these personalities develop peptic ulcers? This is something that we know little about at this time.

Dr. Guin:

In closing, I would like to thank our discussants and to leave the thought with you that we can and do see peptic ulcers in infants and children, not only in relation to those children with central nervous system disease, but also those young individuals under abnormal stress and tension.

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